EVALUATION OF PROGNOSTIC FACTORS IN POSTERIOR URETHRAL VALVE

Dissertation submitted to

Coimbatore Medical College & Hospital,

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For

M.Ch. – Paediatric Surgery

Branch V

The Tamil Nadu

Dr. M. G. R. Medical University

Chennai

AUGUST – 2011

CERTIFICATE

This is to certify that the dissertation entitled **"EVALUATION OF PROGNOSTIC FACTORS IN POSTERIOR URETHRAL VALVE"** is a bonafide record of the work done by **Dr. M.A.VAITHIYALINGAM,** under my guidance and supervision in the Department of Paediatric Surgery during the period of his Post Graduate study at Coimbatore Medical College, Coimbatore for the degree of M.Ch. Paediatric Surgery (Branch V) from 2008 - 2011.

Professor and Head

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DECLARATION

I solemnly declare that the dissertation titled **"EVALUATION OF PROGNOSTIC FACTORS IN POSTERIOR URETHRAL VALVE"** has been prepared by me.

This is submitted to the **Tamilnadu Dr.M.G.R.Medical University**, Chennai in partial fulfillment of the requirements for the award of M.Ch. Paediatric Surgery (Branch V) to be held in August 2011.

Place: Coimbatore

Date: **Dr. M.A.VAITHIYALINGAM**

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This study would not have seen the light of the day, had not our patients showed the kind co-operation they extended. I sincerely thank them.

CONTENTS

INTRODUCTION

Posterior urethral valve represents the most common cause of congenital obstructive uropathy leading to childhood renal failure. The incidence of posterior urethral valve is approximately 1:5000 to 1:8000 infant males. Challenges faced by children with posterior urethral valve are multiple. Obstruction by valve is the process which involves the entire urinary system. Appropriate clinical suspicion remains the key to diagnosis which is confirmed by standard imaging techniques. The risk of renal compromise and ultimate renal failure is a potential problem for each patient. The outcome may be altered by appropriate intervention, but in most cases the renal development in utero determines the need for eventual dialysis or transplantation.

The prognosis for children with urethral valves is improving and current management is gradually rewriting the historical data. In most modern large series neonatal deaths make upto only 2% to 3% of the series.

Early (prenatal) recognition, control of infection, appropriate and selective surgery recognition of harmful urodynamic abnormalities, modern nephrologic management and eventual dialysis and transplantation all combine to increase survival now to an extent unheard of in the past.

In the past, the treatment of posterior urethral valve was based primarily on the mechanism of obstruction and its relief. The current treatment of this condition is based upon our evolving knowledge of the consequences of bladder outlet obstruction. So, the quality of life in these patients, are very much improving in the recent days.

AIMS OF THE STUDY

- To analyze the various described prognostic factors, in our antenatally diagnosed patients.
- To determine the prognostic factors that predict the outcome of posterior urethral valve patients postnatally.
- To identify the significant of the each individual factor in the long term outcome.

MATERIALS AND METHODS

Study Type	:	Retrospective and prospective study
Study period	:	January 2004 to December 2009
Study centre	:	Coimbatore Medical College and Hospital, Coimbatore – 18.

METHODOLOGY

In our study, we registered 52 cases of posterior urethral valves out of which 7 cases are antenatally diagnosed, 24 cases were in the new born period, 13 cases were between 1 to 12 months of age group, 8 cases were between 1 to 4 years of age group.

The initial diagnosis or suspicion of posterior urethral valve based on prenatal ultrasonagraphy, UTI, or others (dehydration, electrolyte changes, palpable bladder, etc.)

The patient's initial evaluation included renal function, urine culture, urine analysis upper and lower urinary tract ultrasonagraphy and MCUG.

Among the 52 patients, 43 patients were treated by primary cystoscopic valve ablation and the remaining cases underwent urinary diversion either vesicostomy (6 cases) or cutaneous ureterostomy.

All the above patients were evaluated regularly throughout their follow-up and accessing the renal function, urine culture and urine analysis; upper and lower urinary tract ultrasonagraphy, MCUG and DMSA scan. The results were analyzed.

Our follow up protocols are

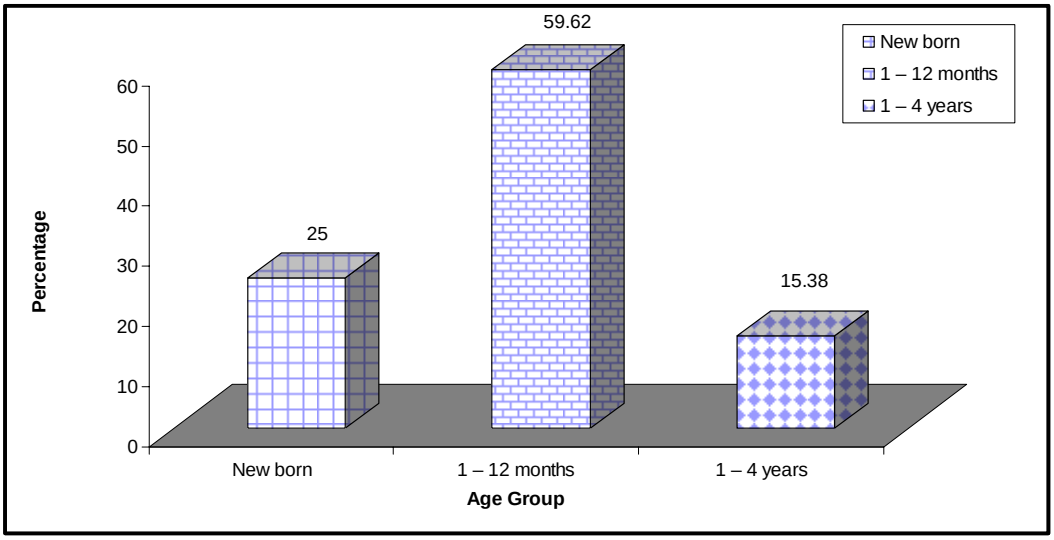
- Accessing the regular stream of urine post operatively,
- Monthly urine for culture and sensitivity
- MCUG after 6 months,
- Ultrasound KUB to assess the upper and lower urinary tract
- DMSA scan in needed patients.

The results were analyzed

RESULTS

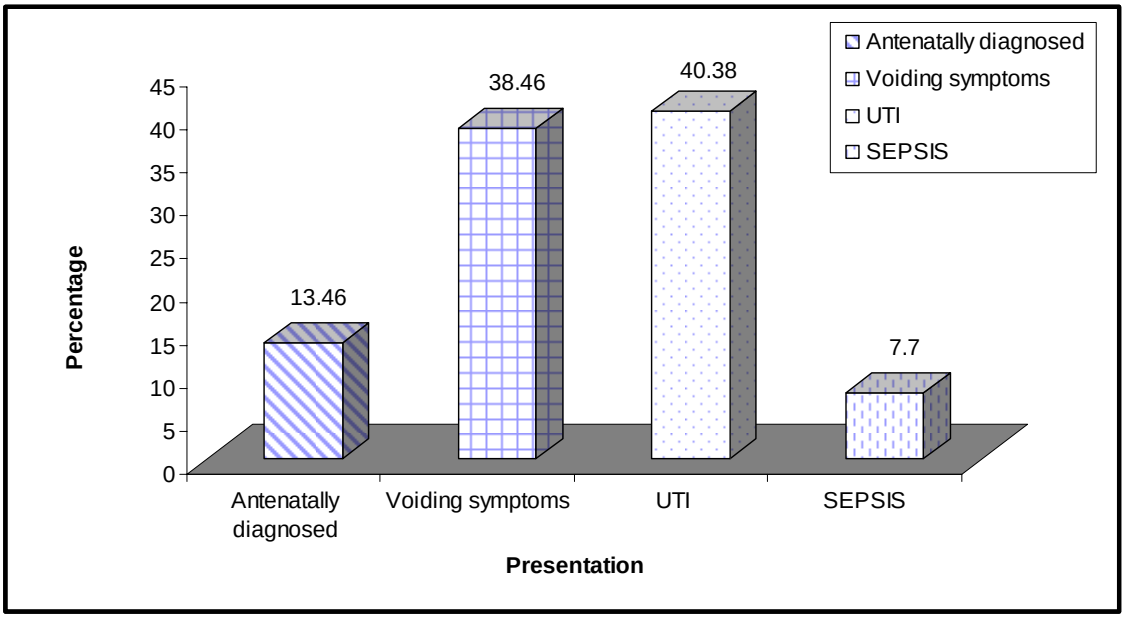
Age distribution

Age Group	No. of patients	Percentage
New born	13	25
1 – 12 months	31	59.62
1 – 4 years	8	15.38
Total	**52**	**100**



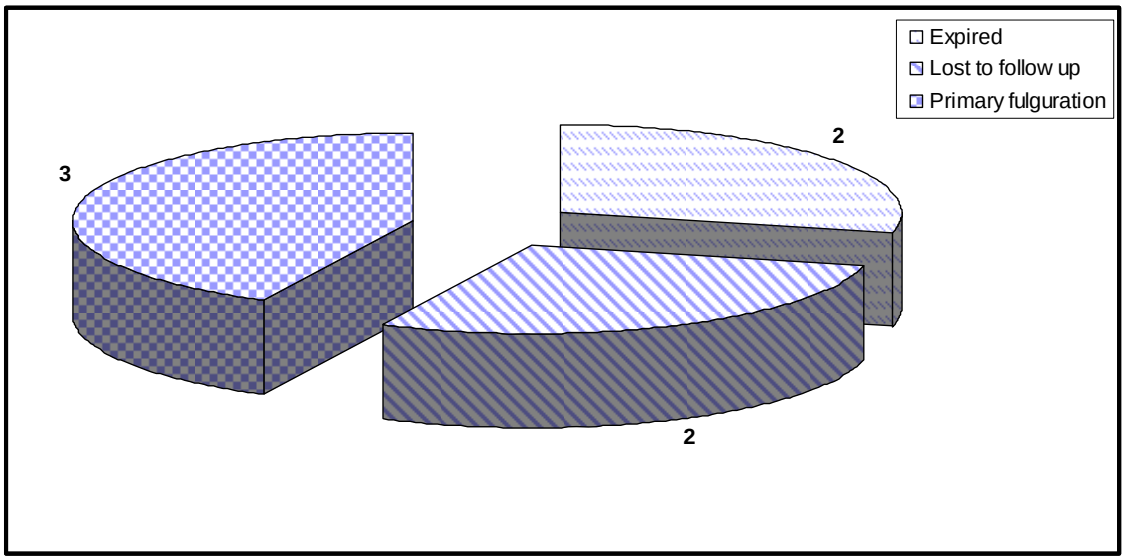
Presentation

Presentation	No. of patients	Percentage
Antenatally diagnosed	7	13.46
Voiding symptoms	20	38.46
UTI	21	40.38
SEPSIS	4	7.70
Total	**52**	**100**



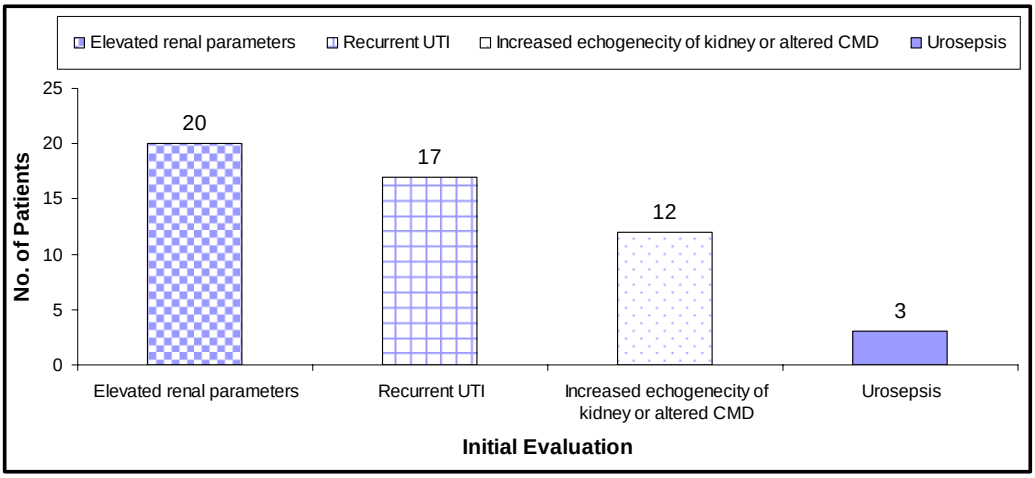
Antenatally diagnosed cases

Expired	2
Lost to follow up	2
Primary fulguration	3



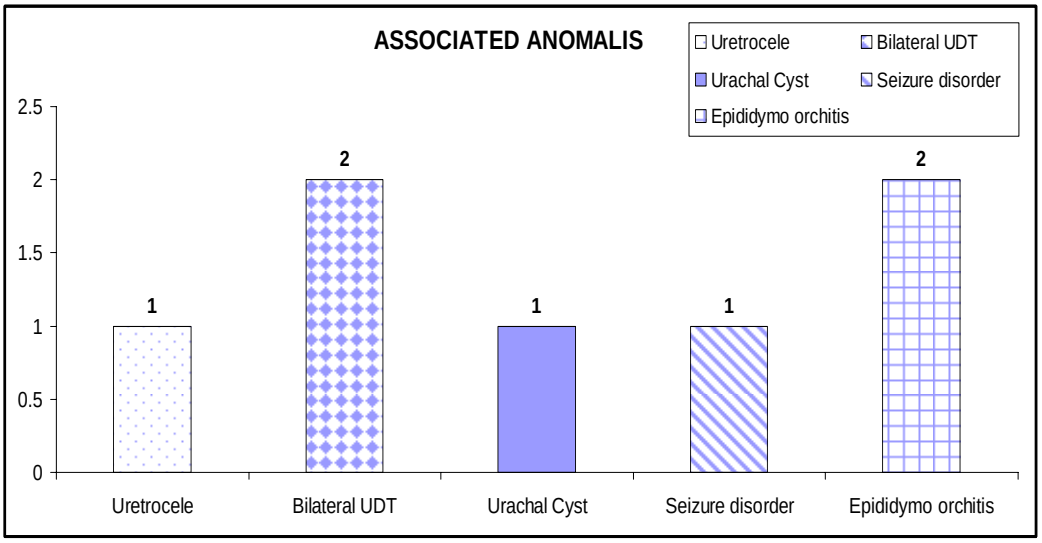
Initial evaluation

Elevated renal parameters	20
Recurrent UTI	17
Increased echogenecity of kidney or altered CMD	12
Urosepsis	3



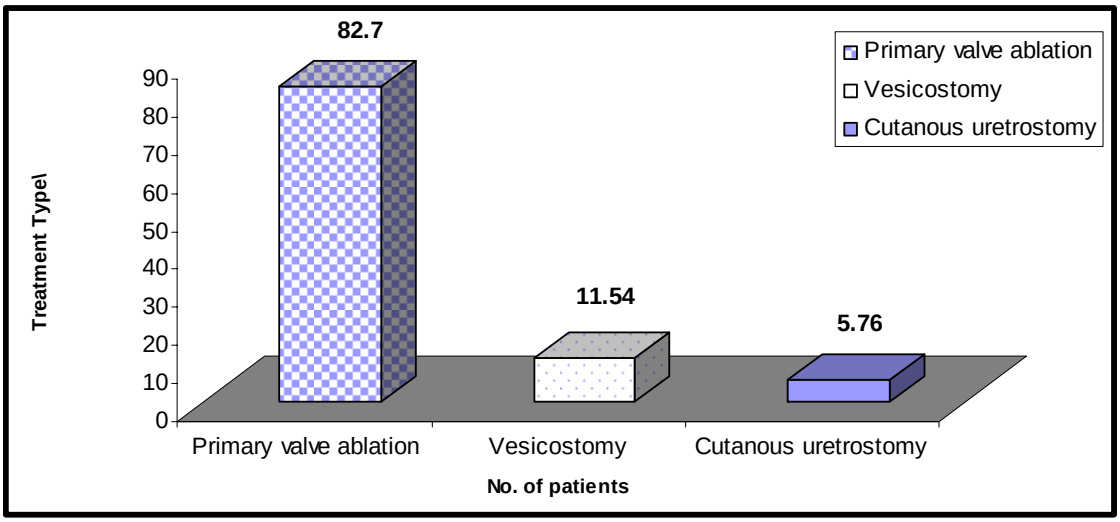
Associated Anomalies

Uretrocele	1
Bilateral UDT	2
Urachal Cyst	1
Seizure disorder	1
Epididymo orchitis	2



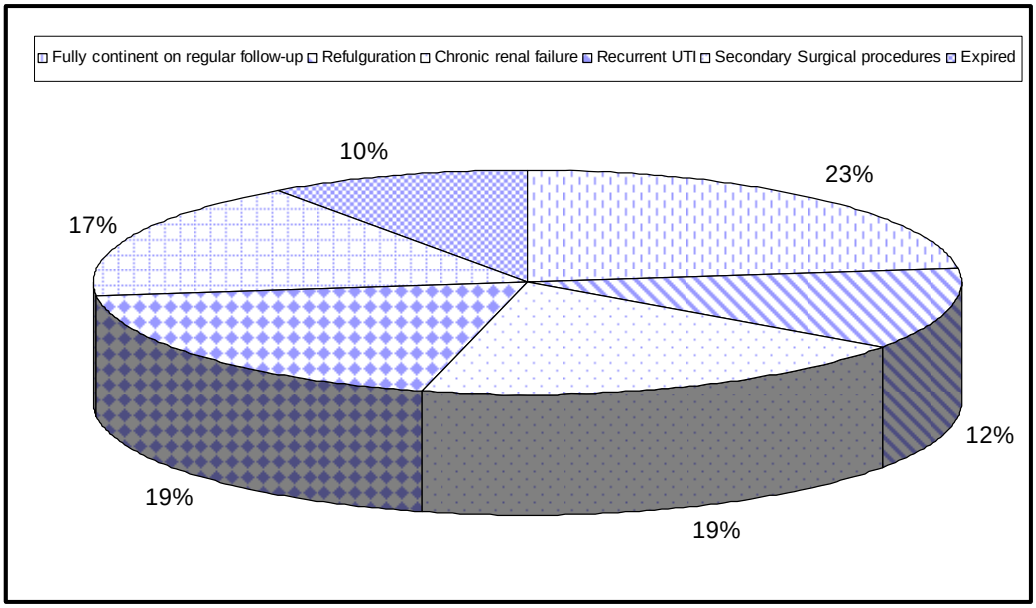
Treatment

Treatment	No. of patients	Percentage
Primary valve ablation	43	82.70
Vesicostomy	6	11.54
Cutanous uretrostomy	3	5.76
Total	**52**	**100**



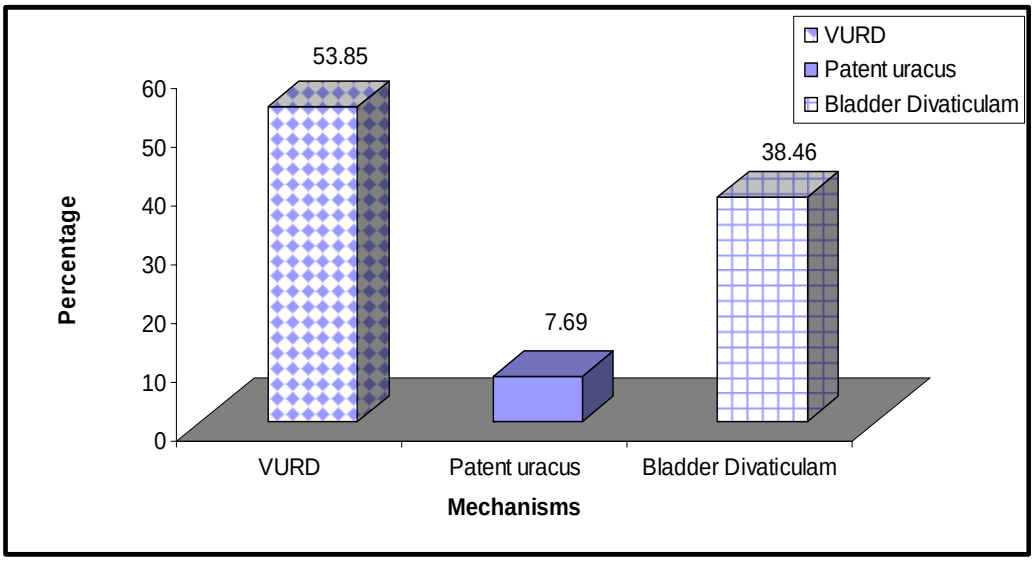
Follow up

Follow up	No. of patients	Percentage
Fully continent on regular follow-up	12	23.08
Refulguration	6	11.53
Chronic renal failure	10	19.23
Recurrent UTI	10	19.23
Secondary Surgical procedures	9	17.31
Expired	5	9.62
Total	**52**	**100**



Pressure Pop Off mechanisms

Mechanisms	No. of patients	Percentage
VURD	7	53.85
Patent uracus	1	7.69
Bladder Divaticulam	5	38.46
Total	**13**	**100**



REVIEW OF LITERATURE

ANATOMY

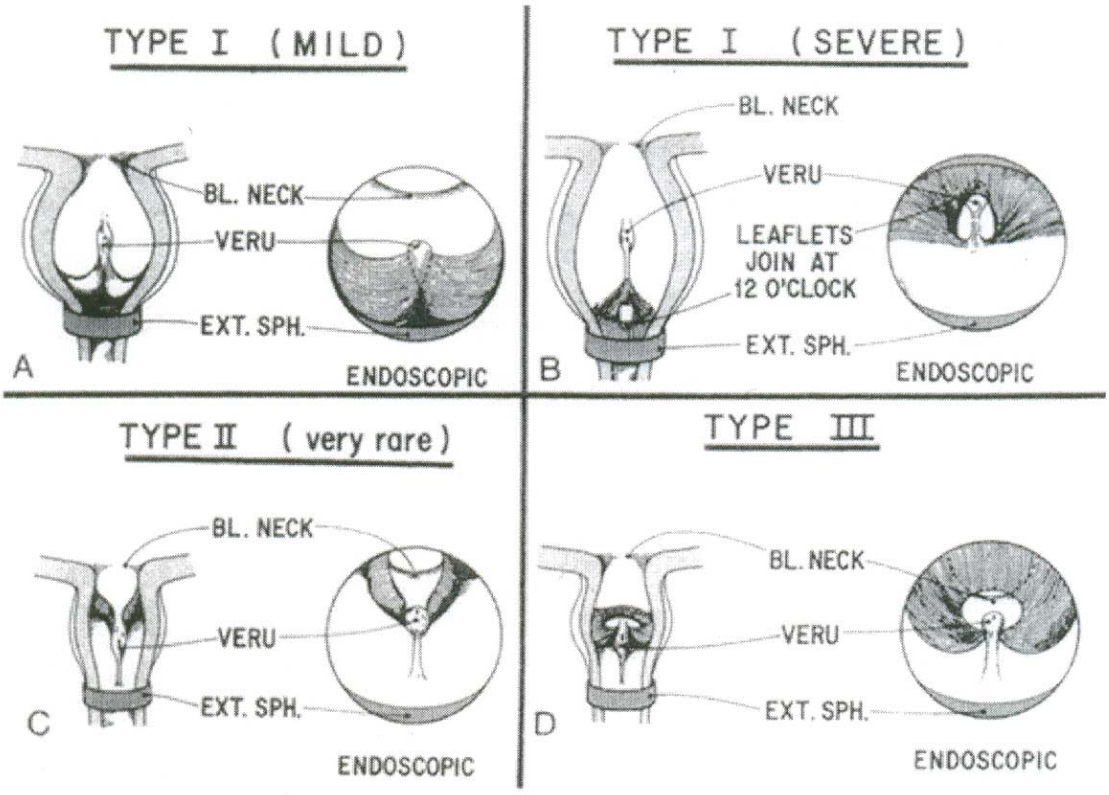
In 1919, Hugh Hampton Young and his associates published their historic description of PUV. He described three type of posterior urethral valve[31].

Young classification is,

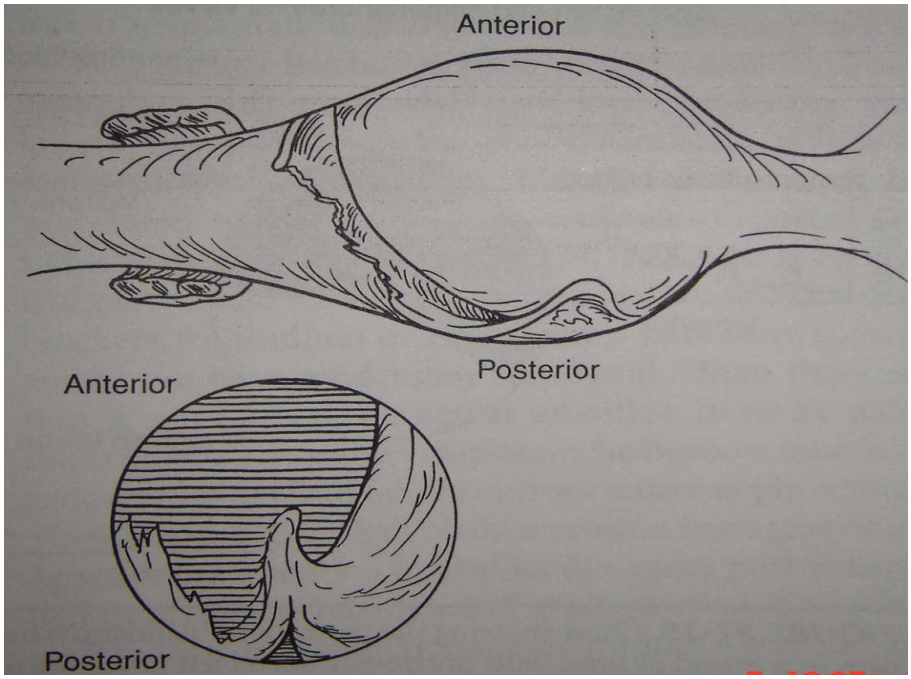
- *Type I valves,* which lie as fins of mucosal tissue that radiate from the urethral crest of the distal verumontanum and sweep across the urethral lumen to fuse anteriorly.
- *Type III valves* are obstructing diaphragms that lie in a transverse plane to the urethral lumen and originate distal to the verumontanum, near the bulbomembranous junction.
- *Type II valves,* rarely mentioned in the literature, are mucosal folds that radiate from the proximal aspect of the verumontanum and extend cephalad to the bladder neck.

In an attempt to provide a simple yet anatomically correct nomenclature for these lesions, Dewan and coworkers advocated replacing the complex Young classification with the unifying

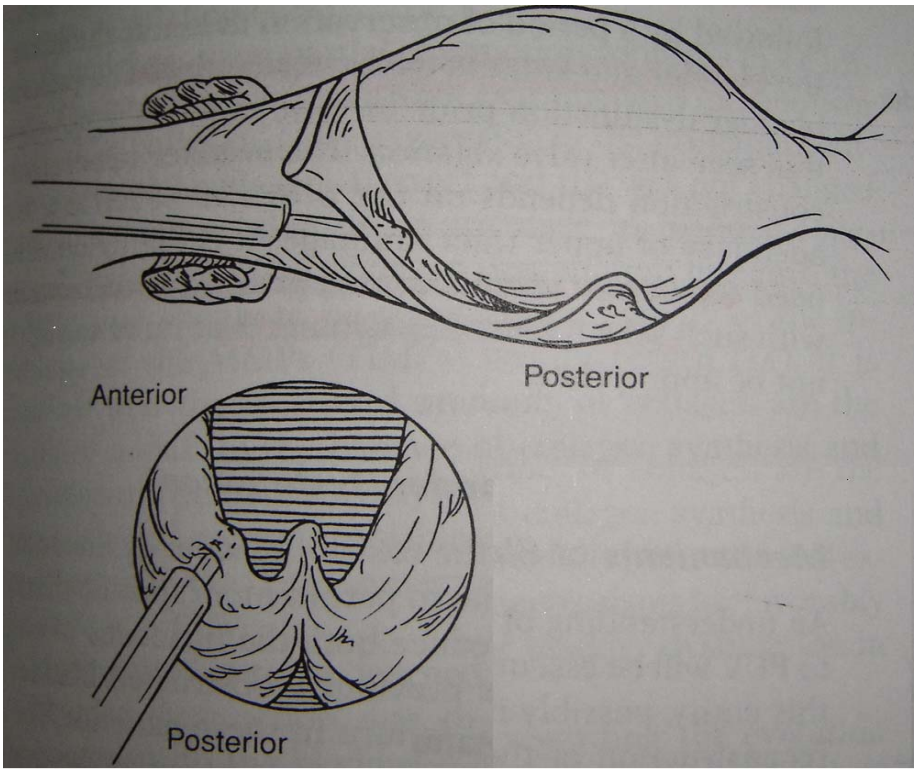
terminology ***congenital obstructing posterior urethral membranes*** **(COPUM).**



POSTERIOR URETHRAL VALVES



PRIMARY VALVE ABLATION

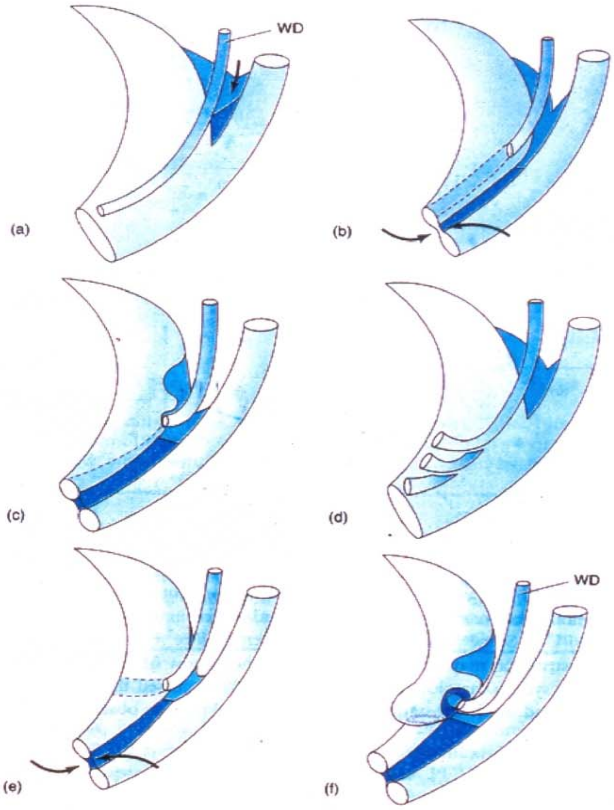


ETIOLOGY

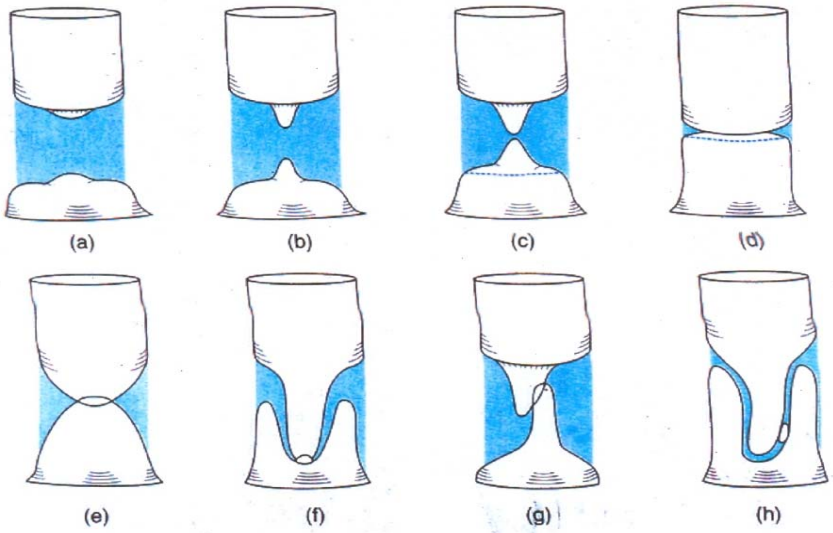
The exact etiology of PUV or membranes has never been established[2]. The prostatic urethra develops from the urogenital sinus by the 8th week of gestational life. The mesonephric ducts and paramesonephric ducts are both absorbed in this region of the vesicourethral canal. The mesonephric ducts develop into the ductus deferens, with the openings (ejaculatory ducts) lying lateral to the verumontanum. The distal paramesonephric ducts form part of the prostatic utricle, a small diverticulum on the verumontanum. The colliculus seminalis or verumontanum forms on the dorsal floor of the prostatic urethra as a result of the elevation of the urethral wall by the expanding ejaculatory ducts and utricle. The ends of the wolffian ducts form the normal urethral crest as they migrate cranially from an anterolateral position in the internal cloaca to a posterior position at the verumontanum.

Stephens and associates hypothesized[3] that the wolffian duct orifices in valve patients are initially misplaced and integrate abnormally into the urethral wall to form the obstructing lesion.

DEVELOPMENT OF TYPE – 1 VALVE



DEVELOPMENT OF TYPE – 3 VALVE



RENAL PATHOLOGY AND URETHRAL VALVES

Renal insufficiency associated with congenital urethral obstruction may result from either primary renal dysplasia or progressive renal deterioration after birth. Experimental models and clinical cases demonstrate the pathogenesis of renal dysplasia associated with urethral obstruction. Severe obstruction in utero has been theorized to transmit damaging back-pressure to the upper tracts, thereby causing deformation of the developing nephrons. Animal data suggest that *early* obstruction may result in severe upper tract changes[4].

Fetal lamb studies showed that early second-trimester ureteral obstruction resulted in renal dysplasia similar to that seen with severe PUV. Similarly, Beck's experimental work in fetal sheep demonstrated dysplasia occurring with early obstruction, whereas later in utero obstruction resulted only in hydronephrosis.

Henneberry and Stephens[5] supported a competing hypothesis known as the "bud theory" of renal dysplasia. They suggested that aberrant caudal budding of the ureter from the

mesonephric duct causes aberrant induction of the renal mesenchyme. In support of this theory[6], they examined 34 renal units from autopsies of valve patients, with 14 of the 19 patients being younger than 6 months of age. They found a significant positive correlation between lateral trigone placement of the ureteral orifice and the gross renal morphologic changes of hydronephrosis and parenchymal thinning.

Histologic evaluation demonstrated lower mean glomerular counts and the most severe degree of dysplasia in renal units with the most lateral ureteral placement. Nevertheless, four renal units with grossly dilated and tortuous ureters demonstrated normal parenchymal development[7]. This finding suggests that the obstruction in these cases may have occurred later in gestation and provides evidence that backpressure and VUR alone are not responsible for dysplasia.

The observation of poor function in the refluxing unit of patients with PUV seems to be very common, more so than in patients who have high-grade reflux without urethral obstruction. This may indicate overlapping pathologies, namely that in utero

pressure work combined with primary dysmorphism results in dysplasia and poor function. There are limited clinical histopathologic data in the literature demonstrating the renal damage associated with urethral obstruction.

A series of renal biopsies[8] from valve patients with renal insufficiency and reflux demonstrated histologic changes of obstruction in 60%, dysplasia in 25%, interstitial fibrosis in 25%, and infectious change in 15%.The authors contended that the relatively low incidence of primary dysplasia in their series provided support to the practice of supravesical urinary diversion. However, the patients studied were not newborns, so these findings may reflect secondary or developmental influences.

In contrast, Tietjen[8] and associates reported renal dysplasia in 85% of renal units from babies treated by proximal diversion for renal insufficiency at birth. Daikha-Dahmane[9] and co-workers analyzed kidney lesions from fetuses demonstrating bilateral urinary tract obstruction and surviving 14 to 37 weeks' gestation. All fetuses older than 20 weeks' gestation showed renal dysplasia with blastema cells, interstitial fibrosis, and an arrest of

nephrogenesis. These findings support the irreversibility of obstructive changes occurring early in gestation.

PRESSURE POP-OFF MECHANISMS IN URETHRAL OBSTRUCTION

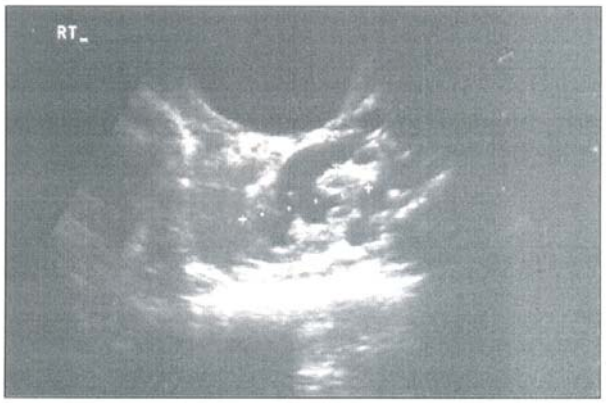
Hoover and Duckett introduced the concept of pressure popoffs in the obstructed urinary tract in 1982. They noted preserved contralateral renal function in patients with unilateral reflux into a nonfunctioning kidney. This phenomenon, known as the valves, unilateral reflux, and renal dysplasia (VURD) syndrome, suggests that mechanisms that relieve bladder pressure may have a protective effect on renal function.

In addition to VUR, other less common pop-off mechanisms include large bladder diverticula, bladder rupture with urinary ascites, and renal urinary extravasation with urinoma formation. There is growing evidence that decompressive mechanisms may also affect bladder development. If the bladder is protected from pressure work during development, can its morphology and function be preserved?[9] Chen and associates presented unusual documentation of three different pop-off mechanisms in a surviving

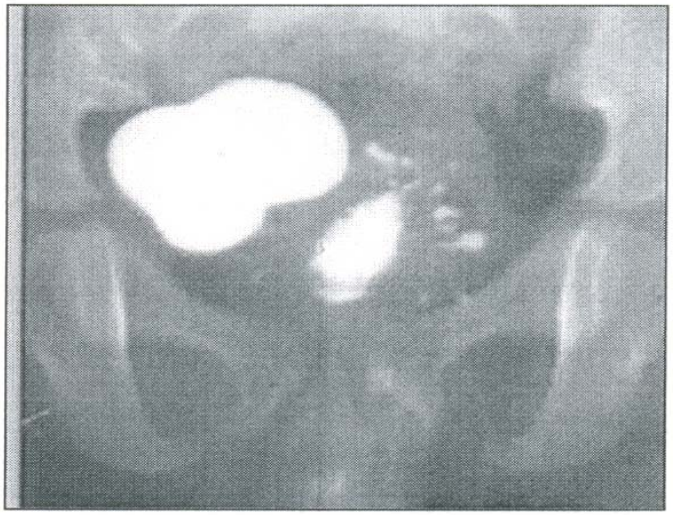
infant with severe PUV. In their patient, oligohydramnios, bladder dome perforation, and ascites were diagnosed at 26 weeks' gestation. At 37 weeks' gestation, bilateral urinomas developed.

Postnatal evaluation revealed bilateral VUR and a small, dystrophic bladder, but there was no long-term follow-up of bladder function. Kaefer[10] and colleagues reported favorable bladder outcomes in 87% of valve patients with pop-off mechanisms. Rather than the typical thick-walled, trabeculated bladder, patients with upper tract pop-off mechanisms may demonstrate a smoothwalled bladder on voiding cystourethrography (VCUG).

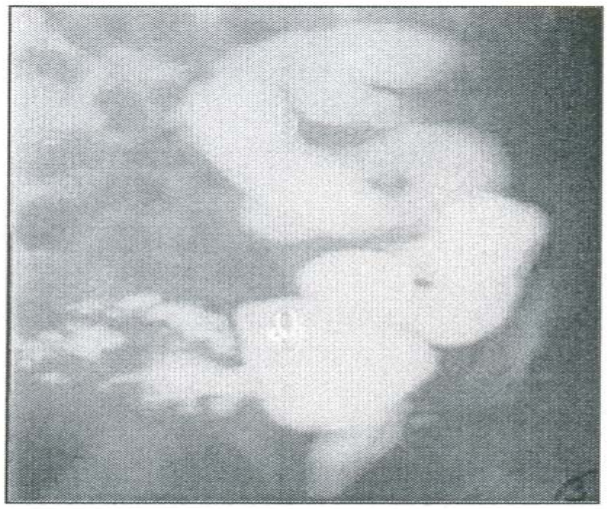
NATURE'S PRESSURE POP- OFF MECHANISMS



PERIRENAL URINOMA



BLADDER DIVERTICULUM



UNILATERAL VUR

PRENATAL DIAGNOSIS OF URETHRAL OBSTRUCTION

In the developed countries where obstetric sonography is routinely performed, most fetuses with urinary tract dilation are detected prenatally. The sensitivity for detection of obstructive

uropathy by prenatal ultrasonography in multiple series was between 90% and 100%.[11] Prenatal ultrasonography should specifically address renal pelvic anteroposterior diameter, amniotic fluid volume, renal echogenicity, renal cortex thickness, bladder distention, bladder wall thickness, presence of urethral dilation, and evidence of urachal patency.

Fetal genitourinary tract screening by ultrasound is possible at 20 weeks' gestation, at which time the kidney is of adequate size for evaluation.[12] A renal pelvic diameter between 4 and 10 mm in the second trimester is considered to represent mild dilation and does not persist postnatally in 97% of cases. A diameter exceeding 10 mm, or the presence of dilated calyces with a pelvic diameter of less than 10 mm, suggests significant pathology and warrants postnatal follow-up.

The fetal renal parenchyma should be evaluated for thickness and echogenicity. Renal cortical echogenicity equal to that of the adjacent liver can occur in infants less than 4 months of age with normal kidneys. Echogenicity brighter than liver or spleen denotes underlying renal pathology. Cortical cysts in a brightly

echogenic kidney are an unmistakable indication of primary renal dysplasia. Cortical atrophy can be seen in association with hydronephrosis and is defined as fetal renal cortex less than 2 mm in thickness. The fetal bladder is more difficult to assess by ultrasonography.[13]

By definition, the bladder wall is considered thickened if it is visible when the bladder is full. Amniotic fluid volume is a key feature of the fetal ultrasound study. If hydroureteronephrosis and persistent bladder dilation are found together, the diagnosis of PUV is suspected, although similar findings may be found in patients with prune-belly syndrome, primary megaureter, or VUR. Of fetuses monitored for persistent megacystis and hydronephrosis suggestive for valves, 42% to 48% had confirmed posterior urethral obstruction at postnatal diagnosis.[14] The findings of increased renal echogenicity and oligohydramnios in addition to hydronephrosis and bladder dilation greatly increase the predictive value of prenatal ultrasonography.

POSTNATAL DIAGNOSIS OF URETHRAL OBSTRUCTION

All newborn males with a history of significant prenatal hydronephrosis should be evaluated for possible urethral obstruction. Because volume depletion is common in the first 48 hours of life, ultrasound studies obtained during this period can be falsely negative and should be repeated at 1 week of age.

The more severe cases of obstruction typically show hydroureteronephrosis, even with relative volume depletion.

A common misconception is that normal voiding guarantees a normal urethra.[15] Although some infants with obstruction have palpable bladder distention or delayed voiding, others have a normal physical examination and regularly wet diapers. A VCUG should be considered before discharge from the nursery if any significant degree of renal collecting system dilation is present on prenatal ultrasonography. In the infant without prenatal renal imaging, severe urethral obstruction can manifest as abdominal distention from a large full bladder, massive hydroureteronephrosis, or urinary ascites. Patent urachus and retroperitoneal urinoma are other findings that raise the suspicion of bladder outlet obstruction.

Newborns with bladder outlet obstruction also present with respiratory distress secondary to pulmonary hypoplasia, severe abdominal distention, or pneumothorax. Other infants with PUV may have a delayed diagnosis made after workup for urinary tract infection, sepsis, acute renal failure, or failure to thrive in the first months of life. The VCUG remains the “gold standard” for postnatal diagnosis of PUV. Transperitoneal ultrasonography is reported to be an alternative diagnostic tool that can demonstrate the urethral changes seen with PUV.

TREATMENT IN THE NEWBORN

The initial treatment in a newborn diagnosed with PUV should begin with bladder drainage by urethral catheter, antibiotic administration, and correction of fluid and electrolyte abnormalities. Creatinine measurements during the first days of life reflect the maternal levels and are not indicative of the infant’s renal function. Serial creatinine measurements taken 7 to 10 days after bladder drainage establish the newborn preoperative nadir in full-term infants.

After this initial period of stabilization, most infants can safely undergo primary ablation of valves. Endoscopic Valve Ablation in the Newborn New pediatric endoscopic equipment has drastically changed the surgical approach to valve treatment. The 8.5F resectoscope with a 5-degree lens and cold knife hook working element can be employed in infants as small as 2000 g.37 Although some surgeons continue to prefer to incise the valves with a Bugbee ball–tipped electrode.

A Bugbee electrode passed through the working channel of a 5F cystourethroscope is a useful technique in very small infants whose urethras cannot accommodate the larger instruments. [24] Potassium-titanyl-phosphate (KTP) laser valve ablation has been reported to be safe in newborn infants, with no urethral stricture formation at 3 years' follow-up; incontinence was not addressed in this study.

Management after Valve Ablation

A common concern of pediatricians managing the patient with PUV is the persistence of severe hydroureteronephrosis after valve ablation.[17] Frequently, this is interpreted as evidence of ureterovesical junction obstruction, and, when it is combined with elevated creatinine levels, it leads to supravesical diversion in some institutions. Chronic dilation of the collecting system and ureters from in utero obstruction or reflux does not resolve immediately but is usually not evidence of continued high intrarenal pressure.

Tietjen18 and associates used the Whitaker test to demonstrate fixed ureterovesical junction obstruction in only 4% of renal units in valve patients who had undergone proximal urinary diversion for newborn renal insufficiency. Furthermore, primary renal dysplasia was identified by biopsy in 85% of these patients. The fact that more than 40% of these patients progressed to end-stage renal failure with proximal diversion supports the belief that renal insufficiency demonstrated after newborn creatinine level stabilization is caused by underlying primary renal dysplasia and is not the result of continued obstruction.

AUGMENTATION (URETEROCYSTOPLASTY)
WITH MITROFANOFF PROCEDURE

NIGHT DRAINAGE

Vigilant follow-up[18] is essential in the neonatal period after hospital discharge. Persistent hydroureteronephrosis is common but should be closely monitored. Urodynamic evaluation can confirm that resting bladder pressures are in a safe range (<30 cm H_2O) and are not the cause of persistent hydronephrosis. Ultrasonography is also used to evaluate the renal parenchymal echogenicity and to demonstrate the presence of corticomedullary junctions. Hulbert and colleagues[19] found that distinct corticomedullary differentiation in infants with PUV imaged before 6 months of age reliably predicted serum creatinine levels lower than 0.8 mg/dl at follow-up 1 to 4 years later.

Although the obstructive effects on the prostatic urethra and bladder neck do not resolve immediately, a postoperative VCUG should confirm complete valve ablation. In cases of severe preoperative urethral distortion, some surgeons prefer to perform repeat cystourethroscopy to rule out and treat possible residual valve tissue.[20]

A nuclear medicine renal scan is best obtained after 4 weeks of life to establish a baseline for differential renal function and to

identify nonfunctioning renal units. Cooperative management by pediatric urology and nephrology staff facilitates the medical management in these infants. Acidosis and salt-wasting nephropathy are common and necessitate frequent monitoring of serum electrolytes.

Infants suffering the most severe degrees of obstruction typically do not survive in utero because of elective termination or fetal demise. If these infants do survive, the degree of renal dysplasia and bladder damage can be so severe that management must be tailored to allow their survival, with acceptance that both renal and bladder function are often unsalvageable.

VESICOURETERAL REFLUX RESOLUTION AFTER PRIMARY VALVE ABLATION

VUR occurs in up to 75% of infants diagnosed with PUV in the first year of life. Traditionally, routine management of these patients has included ureteral reimplantation, often done at the same time as contralateral nephroureterectomy of nonfunctioning renal units. There is long-standing evidence that reimplantation is not necessary in most valve patients, and conservative

management alone is safe and effective. In a series of infants treated by primary valve ablation alone, Close and coworkers found improvement in reflux grade in 18 of 19 refluxing renal units after 1 year.

Complete resolution of reflux occurred within 2 years after valve ablation in 12 (86%) of 14 patients.[22] Reflux into nonfunctioning kidneys typically does not resolve. Nephroureterectomy has been performed in the past to improve voiding dynamics in these patients, although ureteral preservation for possible ureterocystoplasty is appropriate in some cases.

BLADDER FUNCTION IN VALVE PATIENTS

There is wide acceptance of the concept that maximizing the long-term outcome in PUV patients centers on maintaining bladder function. Older patients with a missed diagnosis of severe PUV and valve patients with a history of urinary diversion were described by Mitchell in 1986 as illustrative cases of the “valve bladder syndrome.” [23, 24]

For many, recognition of this syndrome has changed the approach to management of PUV in infants. Boys with a valve bladder demonstrate the long-term effect of persistent renal obstruction from high bladder pressures after relief of urethral obstruction. Pathologic changes in the entire urinary tract combine to further compromise renal function.

Progressive hydroureteronephrosis, polydipsia, polyuria, urinary frequency, and enuresis with renal insufficiency are hallmarks of the syndrome. Renal tubular dysfunction results in a severe urine concentrating defect with polyuria and polydipsia. Urine production in these children can range from 3 to 6 L/day. In patients with VUR and grossly dilated upper tracts, ureteral peristalsis is poor, and large urine volumes result in incomplete emptying of the collecting system, ureters, and bladder. The thick-walled valve bladder is poorly compliant and functionally lacks normal sensation. These patients learn to tolerate high intravesical pressures and are able to hold large urine volumes at these pressures without pain. With gross distention, the thick wall of the bladder causes increased resistance to urine flow through the ureterovesical junction.

Urine holding thus results in increased upper tract dilation and pressure and, ultimately, causes progressive renal damage. There is an increasing focus on the role of the bladder in the long-term outcome of patients with PUV. Urodynamic patterns described in older valve patients include bladder hyperreflexia, hypertonia (noncompliance), and myogenic failure.

Although these changes are attributed to bladder outlet obstruction, the contribution of primary treatment to dysfunction cannot be ascertained from such reports. Many of the studies of bladder and ureteral function in valve patients are complicated by the inclusion of different primary treatment modalities and treatment ages in the same study group. Studies have now been published that focus on the functional outcome in newborns undergoing primary ablation alone for the treatment of congenital urethral obstruction. Holmdahl and associates demonstrated normal bladder compliance by 1 year of age in all infants undergoing primary valve ablation at a mean age of less than 2 months. They did not find the three dysfunctional patterns described by Peters and colleagues in older boys.

In a retrospective evaluation of 23 newborn infants treated by valve ablation in the first weeks and followed up from 1 to 9 years, Close and coworkers[26, 27 & 28] found good bladder function as well as resolution of upper tract abnormalities. VCUG performed 1 year after valve resection showed resolution of trabeculation in more than 85% of patients. Additionally, when compared with infants treated by urinary diversion, those boys undergoing early valve ablation had significantly better bladder compliance and potty-training results.

The mean bladder compliance was 17.2 mL/cm H2O in those infants treated by early primary incision, compared with 5.8 mL/cm H2O in boys treated by diversion. Ninety-two percent of the boys undergoing early ablation were potty trained by 4 years of age, whereas only 17% of the diverted boys were dry by age 4. Low bladder compliance and high bladder pressures led to bladder augmentation in 3 (38%) of the 8 diverted patients. Only 1 (4%) of 23 patients undergoing primary ablation required bladder augmentation, and that patient had severe urethral obstruction that was undetected until 4 months of age.

URETHRAL VALVES AND RENAL TRANSPLANTATION

Chronic renal failure necessitates renal transplantation in a significant number of boys with PUV. With appropriate management, many patients with moderate renal insufficiency at birth reach adolescence before requiring transplantation. Modern studies have addressed the possible detrimental effects of the valve bladder on renal graft survival. Reinberg and colleagues48 demonstrated significantly poorer 5-year graft survival for patients undergoing transplantation for valve-related renal failure than was found in those patients with nonobstructive etiologies. Similarly, Dewan and associates reported that valve bladder led to allograft failure in 12% of valve patients receiving a renal transplant.

Other authors have demonstrated good allograft survival but elevated creatinine levels occurring over long-term follow-up in transplanted valve patients. A large study with 10-year follow-up after renal transplantation demonstrated no difference in graft survival and creatinine levels when comparing children with PUV and children with nonobstructive causes of renal failure. These data may reflect the improvement in urologic management of valve

bladder. The conclusion that the valve bladder will not negatively affect renal allografts is not supported by a long- term follow-up evaluation addressing bladder function and outcomes of renal transplantation. Salomon and coworkers reviewed the voiding history of 44 valve patients who were monitored for a mean of 9 years after renal transplantation.

They found an elevation of serum creatinine after 5 years of follow-up in boys with symptoms of bladder dysfunction including incontinence, urinary urgency, frequency, and difficulty emptying. Because of the relentless effects of the bladder on the upper tracts, the preservation of bladder function must be of primary consideration in all patients with PUV as management decisions are made.

DISCUSSION

Our study included 52 patients of posterior urethral valves. Out of the 52 patients, 13 patients were in the new born period (25%) and 25 patients were infants (59%). The duration of follow ranged from 1 year to 5 years.

On comparing our study with same Cohort of study group by All India Institute of Medical Sciences, Delhi (M.Bajpai et al), the most common age group at age group at initial presentation is in infants.

Among the 38 patients with PUV, elevated serum creatinine value i.e., more than 0.8 mg/dl was present in 10 infants (34%) whereas in new born 6 of 13 babies had elevated serum creatinine level. In the 1 to 4 years of age group, elevated value is present in 4 out of 8 cases.

The most common initial procedure after stabilization of patients with posterior urethral valve was primary cystoscopic

valve ablation which was carried out in 82% of patients. (43 cases).

The remaining patients underwent diversion procedures either vesicostomy (6 patients) or cutaneous uretrostomy (3 patients). Although we have preformed more number of diversion procedures before 2004, with the availability of new born cystoscope we presently perform primary valve ablation in all cases except in few patients.

In few centers like AIIMS, are started doing primary laser vaporization of valves even in the neonatal period but we are not having that facility at present.

The percentage of patients who underwent primary valve ablation was highest in the new born period. (80%). Out of the 52 patients, we did primary valve ablation in 43 cases.

If the patient is not fit for the primary valve ablation either due to urosepsis or poor general condition we went for diversion procedures. 9 cases underwent diversion procedures either vesicostomy or bilateral uretrostomy. Among the 9 patients, 3 were below 1 year of age group, and rests of the patients were above 1

year of age group. Considering this scenario, primary valve ablation is ideal, if the diagnosis of PUV at the earlier age group.

On the initial evaluation, 20 out of 52 patients, had elevated f serum creatinine (>1.2 mg/dl) level. Out of the 20 patients, 10 had normal creatinine level i.e., <0.8 mg/dl at the age of 12 months and maintain within <1 mg/dl in the followup period. 2 patients had expired due to urosepsis and chronic renal failure. 8 patients had renal insufficiency in the regular follow up. Among the 8 patients, 4 of them had vesico uretric reflux and renal scars in DMSA scan.

Considering this, serum creatinine value at the initial evaluation, at the age of 1 year and the final follow up in an individual prognostic factor that determines the outcome of the PUV in our group.

In this observation most of the patients in neonatal age group had renal failure at initial presentation, but one half of them had regained normal renal function by the time of followup. In patients with 1 to 4 years of age group, the incidence of renal failure at present was similar (11 out of 21 patients) but the recovery was

not liked early presentation. 7 patients had renal insufficiency in the followup period.

So early age at presentation is another good prognostic indicator in our observation.

Out of 9 patients who underwent diversion procedures initially,6 patients had improved and their renal function regained to normal in 2 years followup , even though they undergone various secondary surgical procedure like bladder augmentation, Mitrofanoff's procedure etc., So the treatment modalities either primary valve ablation or diversion procedure does not affect the outcome of disease process in our study group.

During the initial evaluation with ultra sonogram, 11 patients had sonographically identifiable abnormalities in kidneys and bladder (absence of cortico medullary differentiation, increased echogenecity of kindey and thickend bladder). Out of the 11 patients 7 had chronic renal failure in the follow-up period, 3 of them expired, 1 patient had lost to follow-up, probably expired.

So increased echogenecity of kidneys comparing with adjacent liver or spleen and absence of cartico medullary differentiation is a bad prognostic indicator in our observation.

The presence of pressure Pop Off mechanisms like VURD, Patent urachus, urinary ascitis, bladder diverticuulum are considered to be a good prognostic factors for various texts. But in our study it's not like so. We had 12 patients with pressure pop off mechanisms (7 VURD , 1 patent urachus and 4 bladder diverticulum). Out of the 12 patients 7 had renal insufficiency in the follow ups. It was probably due to late presentation. (All these patients were presented above 1 year)

So the presence of pressure pop off does not alter the long term outcome of disease in our observation.

Presence of vesico uretric reflux (18 patients) also did not affect the long term outcome.

Only few numbers of PUV patients (7 out of 52) diagnosed antenatally in our group. Among the 7 patients we had lost 4

patients. (Either expired or lost to follow-up).Only 3 patients came for treatment and they underwent primary valve ablation. Out of the 3 patients, 1 patient expired in the follow up period.

So the prognosis of antenatally detected cases of PUV in our centre is dismal.

Considering the outcome of primary valve ablation, out of the 43 patients, 6 had residual valves in the follow-up period, who needed residual valve ablation. In the remaining patients urinary stream became normal. All these patients are fully continent in the follow-up.

Seven of our patients needed secondary surgical procedures like bladder augmentation, ureteric reimplantation, ureterocele excision etc., All these patients are in regular follow-up and 3 of them had renal insufficiency.

Presence of 1 or more renal scars in DMSA scan signifies that the kidney would go for a progressive failure later. 7 of the

patients had renal scars initially and all these patients had renal failure on further follow-up.

During the follow-up period of 5 years, 5 of our patients had expired due to chronic renal failure and urosepsis.

Among the 52 patients 12 patients had chronic renal failure and they are now in regular nephrological followp.

Recurrent urinary tract infection had present in 10 of our patients post operatively posing a great challenge to treat and it is mainly due to presence of high grade reflux.

Considering all these observations only 18 of our patients (34%) had good continence and no renal insufficiency in the 5 years follow-up and had a good quality of life.

Various described prognostic factors in the literature

Variable	Good Predictors	Poor Predictors
In – utero presentation (Weeks)	>24	<24
Amniotic fluid volume	Normal to moderately increased	Moderate to severely decreased
Sonographic appearance of renal parenchyma	Normal to slightly increased echogenicity	Increased echogenicity to frankly cystic
Fetal urinary values		
Sodium (mEq/L)	<100	>100
Chloride (mEq/L)	<90	>90
Osmolality (mOsm)	<210	>210
Urinary output (ml/Hr)	>2	<2
Beta 2 microglobulin	<6	>6

Prognostic variables – after birth

Variable	Good Predictors	Poor Predictors
Sonographic identification of CMJ differentiation	Present Pyramids in atleast one kidney	Absent Hyperechoic, no pyramids
S.Crreatinine	< 0.8 at one year	> 0.8 at one year
Reflux	No reflux	Bilateral reflux
Continence	At 5 years	Incontinence
Pop off mechanisms Urinary Ascites Bladder diverticulum VURD] Patent urachus	Present Present Present Present	Absent Absent Absent Absent

In our study, only less number of patients are turned up antenataly so the antenatal assessment of prognosis is difficult. So most of the patients were assessed postnatally with available investigations and clinical examinations.

Indian Scenario of PUV

The incidence of antenatal diagnosis is only 10%in India. Urinary diversion is done in 50% of cases in most centers, in

tertiary centres 80% of cases are ablated primarily. The results of laser fulguration of valves from AIMS, New Delhi and the primary newborn fulguration from other centers in India are encouraging. Abraham from Kerala, Gopal from Varanasi and Kulasekar from Colombo have developed hooks for valve ablation but the use is limited to their own centers only. Fetal surgery is not done in any of our tertiary centers.

CONCLUSION

Early age group (<1 month) at initial presentation is a single most prognostic indicator in our observation.

Serum creatinine level at the time of diagnosis, 12 months after valve ablation and at the time of last followup is the main factor that indicates the outcome of the disease.

The treatment modalities either primary valve ablation or diversion procedures such as vesicostomy or cutaneous ureterostomy does not affect the outcome of disease process.

The prognosis of our antenatally detected PUV patients were dismal

Absence of corticomedullary differentiation and altered cortical echos in ultrasonography predict the poor prognostic outcome.

Presence of vesicoureteric reflux does not have any impact in the long term outcome.

Presence of pressure pop of does not have any significance in our observation.

Presence of one or more renal scars in DMSA scan is a definitive predictor for future renal impairment.

Only 34% of our patients had good quality of life without renal insufficiency in the 5 year follow-up.

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PROFORMA

POSTERIOR URETHRAL VALVE – FOLLOW - UP

Name	Age
IP No.	PS No.
Wt	Height
Age at diagnosis	Age at surgical internvetion
Antenatal USG	
Urine albumin	
Urine Culture	
Bl Urea	Sr.Creatinine
Sr Electrolytes	Na^+ K^+
Postnatal Ultrasound	
MCU – Bladder	
Reflux Rt	
Lt	
Primary Procedure	
Post op stream – good/fair/poor	
P/a Bladder	

FOLLOW UP

Wt		
Urine alb		
Sp. Gravity		
Sr Electrolytes	Na^+	K^+
Bl Urea	Sr Cretinine	
Urine culture		
MCU		
Ultrasound		
Crystoscopy		
Urodynamics		
DMSA		
Procedure		

MASTER CHART

Sl. No.	Name	Age	Serum creatinine Level at Initial presentation	Treatment
1	B/o Chitra	17 days	<0.8	Primary valve ablation
2	Harish	3 months	2.1	Primary valve ablation
3	Lavan Kishore	2 months	1.4	Primary valve ablation
4	B/o Kalaiselvi	14 days	0.8	Primary valve ablation
5	Kingisly Josuva	4.5 months	0.7	Primary valve ablation
6	Sathish	7 months	1.6	Primary valve ablation
7	Hariharan	1/2 year	2.2	Primary valve ablation
8	Ashok Kumar	8 months	0.7	Primary valve ablation
9	Dinesh	9 months	0.8	Primary valve ablation
10	Lingamurthy	2 year	1.7	Diversion
11	Akash	2 year	2	Primary valve ablation
12	B/o Saraswathy	22 days	0.6	Primary valve ablation
13	B/o Nagaswari	26 days	2	Primary valve ablation
14	Vinnarasan	10 months	2.2	Diversion
15	B/o Ramanathan	15 days	0.8	Primary valve ablation
16	B/o Megala	3 months	0.7	Primary valve ablation
17	B/o Divya	12 days	0.6	Primary valve ablation
18	B/o Ranjitha	2 months	1.8	Primary valve ablation
19	Nandakumar	2 year	0.8	Diversion
20	Gopalakrishnan	4 year	2.3	Diversion
21	B/o Shanthi	1.5 months	0.8	Primary valve ablation
22	B/o Kavitha	19 days	0.6	Diversion
23	B/o Sangeetha	4 months	0.7	Primary valve ablation
24	B/o Mariyammal	3 months	0.8	Primary valve ablation
25	Thoufiq	12 months	0.7	Primary valve ablation
26	B/o Sumathi	7 months	0.9	Primary valve ablation

Sl. No.	Name	Age	Serum creatinine Level at Initial presentation	Treatment
27	Sakthivel	11 months	0.8	Primary valve ablation
28	B/o Sudha	6 months	0.7	Primary valve ablation
29	Ranjith Kumar	2 year	1	Primary valve ablation
30	B/o Saranya	12 days	0.6	Primary valve ablation
31	B/o Vanitha	10 days	0.7	Primary valve ablation
32	Prasanna	3 months	1.5	Diversion
33	Johnson	7 months	0.7	Primary valve ablation
34	Joas	2 year	2	Primary valve ablation
35	B/o Kalyani	24 days	0.7	Diversion
36	B/o Santhiya	7 days	0.8	Primary valve ablation
37	Atheswaran	1.5 year	1.6	Primary valve ablation
38	Mohamed Thoha	7 months	0.7	Primary valve ablation
39	Sanjai	8 months	0.6	Primary valve ablation
40	B/o Chitra	4.5 months	1.6	Primary valve ablation
41	B/o Chandra	6 months	0.6	Primary valve ablation
42	B/o Divya	6 days	0.8	Primary valve ablation
43	Hariharan	1.5 year	1.1	Diversion
44	B/o Mariyammal	27 days	1.6	Primary valve ablation
45	B/o Anandi	3 months	1.2	Primary valve ablation
46	Rama Krishnan	2 year	1	Diversion
47	Santhosh Kumar	11 months	0.7	Primary valve ablation
48	Adithiya	12 months	<0.8	Primary valve ablation
49	Akilesh	3 year	2	Primary valve ablation
50	Rajeswaran	13 year	1.8	Primary valve ablation
51	Kalaiyesan	2 year	1.5	Primary valve ablation
52	Vasanth	2.5 year	1.2	Primary valve ablation